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The Challenging Clinical Management of Patients with Cranial Dural Arteriovenous Fistula and Secondary Parkinson's Syndrome: Pathophysiology and Treatment Options

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Original Paper

The Challenging Clinical Management of Patients with Cranial Dural Arteriovenous Fistula and Secondary Parkinson's Syndrome: Pathophysiology and Treatment Options

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Keywords

Intracranial dural arteriovenous fistula · Venous sinus thrombosis · Shunt · Venous hypertension · Parkinsonism

Abstract

Cranial dural arteriovenous fistula (cDAVF) may rarely lead to parkinsonism and rapid cognitive decline. Dysfunction of the extrapyramidal system and the thalamus, due to venous congestion of the Galenic system with subsequent parenchymal edema, is likely to represent an important pathophysiological mechanism. Here, we report a case of a 57-year-old man with a cDAVF of the straight sinus (Borden type III; DES-Zurich bridging vein shunt [BVS] type with direct, exclusive, and strained leptomeningeal venous drainage [LVD]) and subsequent edema of both thalami, the internal capsule, the hippocampi, the pallidum, and the mesencephalon. Several attempts at venous embolization were unsuccessful, and the neurological condition of the patient further deteriorated with progressive parkinsonism and intermittent episodes of loss of consciousness (KPS 30). A suboccipital mini-craniotomy was performed and the culmenal vein was disconnected from the medial tentorial sinus, achieving an immediate fistula occlusion. Three-month follow-up MRI revealed complete regression of the edema. Clinically, parkinsonism remitted completely, allowing for tapering of dopaminergic medication. His cognition markedly improved in further course. The purpose of this report is to highlight the importance of rapid and complete cDAVF occlusion to reverse venous hypertension and pre-

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vent progressive clinical impairment. The review of the literature underlines the high morbidity and mortality of these patients. Microsurgical disconnection of the fistula plays an important role in the management of these patients and, surprisingly, has not been reported so far.

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Introduction

A cranial dural arteriovenous fistula (cDAVF) is an acquired vascular malformation characterized by arteriovenous (AV) shunting within one of the following venous structures: the dural sinus (dural sinus shunt [DSS]), the transdural segment of the bridging veins (bridging vein shunt [BVS]), or the emissary and epidural veins (emissary vein shunt [EVS]) [1–3]. cDAVF represents approximately 10–15% of all intracranial AV malformations and has been described as being caused by traumatic injuries, venous sinus inflammation, brain surgery, and a hypercoagulable state [4], but the most plausible explanation of its development is sinus thrombosis, mostly as consequence of the previously listed conditions [2, 5].

The clinical manifestation of cDAVF depends on its anatomic location and the pattern of venous drainage. cDAVF with leptomeningeal venous drainage (LVD), also called cortical venous reflux, carries the highest risk of an aggressive clinical course and may present with venous congestion-related parenchymal edema, microbleeds, or massive hemorrhages, and subarachnoid bleeding due to the rupture of venous varices [6]. Borden et al. [4] classified these lesions as type 2 and type 3. Cognard et al. [7] described them as type IIb, IIa + IIb, III, IV, and V. According to the DES (direct, exclusive, strained)-Zurich classification, LVD is an inherent characteristic of all BVS-type fistulae. Due to their specific venous angioarchitecture, DSS-type fistulae and EVS present with LVD frequently and infrequently, respectively [2, 3, 6].

A rare, but severe consequence of cDAVF at a specific location is the occurrence of parkinsonism with/or without progressive cognitive decline. Parkinsonism as a clinical manifestation of cDAVF is exceptionally rare and has only been reported in case reports (Table 1) [8–26]. Dysregulation of the extrapyramidal system and the thalamus due to venous congestion of the Galenic system, with subsequent parenchymal edema, seems to represent an important pathophysiological mechanism here [20].

The early diagnosis and prompt treatment of cDAVF with LVD is mandatory to reverse the venous congestion and eliminate the risk of bleeding [4]. Complete occlusion of the cDAVF is obligatory to achieve neurological recovery and needs to be urgently performed. However, the incidence of incomplete cDAVF occlusion in these patients, which leads to further neurological deterioration, and even death, is surprisingly high in the literature [11, 12, 14, 22, 24, 25].

The rare incidence of cDAVF leading to parkinsonism and cognitive dysfunction and the challenging treatment prompted us to report this case and discuss the current management of cranial cDAVF based on a literature review.

Case Report

A 57-year-old man presented to an external hospital due to rapidly progressive cognitive impairment over 3 months. Neurological examination revealed an altered mental status, with impaired executive function and a hypokinetic movement disorder with gait impairment, hypophonia, and hypomimia. The patient had known reflux esophagitis but no other signif-

Table 1. Overview of published cases with parkinsonism due to cDAVF: Anatomical features of cDAVF and their clinical management

Case No.	First author [ref.], year	Age, yrs /sex	Fistula location	cDAVF etiology – risk factors	cDAVF anatomy (feeding vessels, venous reflux)	cDAVF classification	Neurological symptoms	Time from onset to treatment	Hyperintense lesions on T2-weighted image	Therapeutic intervention	Medication	Follow-up overall outcome
<i>Described cases with cDAVF and secondary parkinsonism</i>												
1	Okuzumi [8], 1998	81/M	R transverse sinus	n.d.	n.d.	n.d.	dementia, parkinsonism and myoclonus of the extremities	6 mo.	bilateral cerebral white matter	endovascular embolization → n.d./no angiography, unclear if complete occlusion was achieved	L-dopa 300 mg daily	improved
2	Matsuda [9], 1999	55/M	R sigmoid sinus	bilateral occlusion of the jugular veins	fed by the R occipital artery and branches of the R MMA; venous reflux to the straight sinus and SSS	n.d.	moderate rigidity and bradykinesia predominant on the L side; dementia	8 mo.	bilateral deep and subcortical white matter	2 series of selective TAE → n.d./no angiography, unclear if complete occlusion was achieved	n.d.	improved
3	Matsuda [9], 1999	78/M	R sigmoid sinus	removal of R jugular vein during pharyngo-esophagectomy; occlusion of the L jugular vein	fed by the R occipital artery and branches of the R MMA; venous reflux to the straight sinus, SSS, and frontal cortical veins	n.d.	moderate rigidity and bradykinesia were found at both extremities; dementia	9 mo.	n.d.	2 series of TAE therapy → n.d./no angiography, unclear if complete occlusion was achieved	n.d.	improved
4	Matsuda [9], 1999	69/F	L sigmoid sinus	L jugular vein occluded, R sigmoid sinus stenosed	fed by the L occipital artery, branches of the L MMA and posterior branch of the L vertebral artery; venous reflux to the L transverse sinus, SSS, and cortical veins	n.d.	marked rigidity and bradykinesia were found on both extremities, but no tremor	some yrs	bilateral cerebral white matter	TAE therapy → n.d./no angiography, unclear if complete occlusion was achieved	n.d.	remained comatose
5	Hamada [19], 2003	44/M	anterior cranial fossa	n.d.	fed by the R anterior ethmoidal artery	n.d.	gait disturbances with “frozen gait”	1 wk	none	n.d.	n.d.	improved
6	Lee [20], 2005	60/F	L TSS	none	fed from the L occipital artery and branches of the MMA; retrograde filling of the DV system and SSS with prominent cortical venous reflux	Cognard grade IIIa+b	bilateral action tremor, severe bradykinesia, facial hypomimia	18 mo.	bilateral subcortical white matter	selective TAE → n.d./no angiography, unclear if complete occlusion was achieved	L-dopa up to 800 mg daily	improved
7	Kajitani [21], 2007	75/M	R TSS and SSS	occlusion of transverse sinus	n.d.	n.d.	bilateral postural tremor	14 wks	none	R occipital artery occluded by TAE, with slight clinical benefit; 1 month later, the patient received feeding artery coagulation surgery and a second session of TAE	none	improved

Table 1 (continued)

Case No.	First author [ref.], year	Age, yrs /sex	Fistula location	cDAVF etiology – risk factors	cDAVF anatomy (feeding vessels, venous reflux)	cDAVF classification	Neurological symptoms	Time from onset to treatment	Hyperintense lesions on T2-weighted image	Therapeutic intervention	Medication	Follow-up overall outcome
8	Miura [22], 2009	65/M	L transverse-sigmoid sinus	L sigmoid sinus narrowed	fed by branches of the left vertebral artery and external carotid artery; retrograde flow into the straight sinus and DV system with cortical reflux	n.d.	parkinsonism and ataxia	11 mo.	basal ganglia	3 series of TAE → incomplete fistula occlusion	n.d.	improved
9	Nogueira [23], 2009	79/M	left transverse-sigmoid sinus	n.d.	supplied by the L occipital middle meningeal (petrosal branch), and posterior auricular arteries, as well as the posterior temporal branches of the L posterior cerebral artery and the lateral clival branch of the cavernous left internal carotid artery; retrograde filling of the SSS and DV system, with severe venous congestion in the posterior fossa	Cognard grade IIa + b	gait imbalance, postural instability, and tremor	2 yrs	n.d.	Step 1: N-butyl-cyanoacrylate and platinum coil embolization Step 2: coil embolization Step 3: combined surgical and endovascular approach with surgical access to the L transverse sinus followed by TV coil embolization of the L sigmoid sinus and lateral aspect of the L transverse sinus. → complete obliteration of the cDAVF was achieved	n.d.	improved
10	Netravathi [24], 2011	54/M	torcula	no visualization of bilateral transverse sinus, straight sinus, vein of Galen and internal cerebral veins suggestive of thrombosis; procoagulant workup revealed protein C and protein S deficiencies	fed by the transosseous meningeal branches of the R occipital artery and the meningeal branches of the L ascending pharyngeal artery, and draining into an irregular straight sinus; retrograde flow into the vein of Galen, cavernous sinus, superior ophthalmic vein, and pterygoid venous plexus	n.d.	hypomimia, postural tremor, generalized bradykinesia and mild rigidity of the upper limbs, brisk stretch reflexes, equivocal plantar reflexes and a positive snout reflex and glabellar tap	3 yrs	bilateral symmetrical thalamic and pallidal edematous lesions, with focal areas of hemorrhage	embolization of the occipital feeding vessels → incomplete occlusion of the cDAVF	n.d.	minimal improvement

Table 1 (continued)

Case No.	First author [ref], year	Age, yrs /sex	Fistula location	cDAVF etiology – risk factors	cDAVF anatomy (feeding vessels, venous reflux)	cDAVF classification	Neurological symptoms	Time from onset to treatment	Hyperintense lesions on T2-weighted image	Therapeutic intervention	Medication	Follow-up overall outcome
11	Netravathi [24], 2011	40/M	SSS	distal R sigmoid sinus and jugular vein were not visualized	multiple feeders from the bilateral internal carotid arteries (tentorial branches), superficial temporal and middle meningeal branches; retrograde sinus flow and cortical venous reflux	n.d.	hypomimia, a reduced blink rate, mild papilloedema, mild rigidity of the upper limbs, generalized bradykinesia, tandem ataxia, hyperreflexia, and bilateral extensor plantar reflexes	3 mo.	multiple hyperdense enhancing lesions in grey and white matter	TV embolization and TAE of the cDAVF was attempted but was unsuccessful; occlusion of the cDAVF was not achieved	L-dopa	further deterioration
12	Hattori [25], 2012	52/F	R transverse-sigmoid sinus	R sigmoid sinus had a proximal stenosis, contralateral transverse sinus was hypoplastic, posterior third of SSS had stenosis	feeding arteries were the MMA, the posterior auricular, occipital, and tentorial arteries; R cerebral blood flow mainly drained to the contralateral sigmoid sinus through the cortical veins; reflux to the straight sinus and cortical veins	n.d.	akinetetic mutism, rigidity, short-stepped gait	3 mo.	basal ganglia and deep white matter of the R occipital lobe	TV coil embolization → first follow-up angiography 1 month later shows disappearance of the TSS fistula, but de novo at the left transverse sinus → embolization → 3 years later de novo cDAVF at SSS → observation was performed → occlusion of the cDAVF was not achieved	"under medication"	improved
13	Shahar [26], 2012	59/M	straight sinus; cDAVF adjacent to right tentorial notch	occlusion of straight sinus	fed by the R MMA (MMA) and a branch of the R occipital meningeal artery; the cortical venous drainage associated with the varicose drained into the vein of Galen, which flowed in a retrograde direction into the vein of the vein of Rosenthal, as well as into the internal cerebral veins	n.d.	rigidity in all 4 limbs, with some hypokinesia on rapid alternating movements; the patient had a slow, hesitant gait with short steps and decreased arm swing bilaterally	1 mo.	basal ganglia	endovascular embolization was performed through the posterior branch of the right MMA → complete occlusion was achieved	n.d.	improved
14	Geraldes [10], 2012	64/M	torcula	thrombosis of distal sagittal and proximal lateral sinuses	torcular dural fistula, with the posterior occipital arteries and R MMA; draining to the straight sinus DV system and marked venous stasis	Djindjian type IIb	progressive cognitive decline, extrapyramidal and cerebellar signs, and myoclonus	3 mo.	T2 basal ganglia hypersignal, and no deep white matter changes	endovascular embolization → complete occlusion was achieved		slightly improved

Table 1 (continued)

Case No.	First author [ref.], year	Age, yrs / sex	Fistula location	cDAVF etiology – risk factors	cDAVF anatomy (feeding vessels, venous reflux)	cDAVF classification	Neurological symptoms	Time from onset to treatment	Hyperintense lesions on T2-weighted image	Therapeutic intervention	Medication	Follow-up overall outcome
15	Jagtap [11], 2014	73/F	DAVF at junction of bilateral transverse sinus-sigmoid sinus	no DSA	hypertrophied feeders from extracranial arteries bilaterally	n.d.	progressive cognitive decline, gait difficulty, and myoclonic jerks	3 mo.	bilateral cerebral hemispheres	none		died
16	Luo [12], 2014	54/M	R TSS	no DSA available	n.d.	n.d.	slowness of movement, cognitive dysfunction, and urinary incontinence	10 mo.	inner part of L temporal lobe	none	Madopar 187.5 mg 4× daily	further neurological deterioration
17	Luo [12], 2014	75/M	L TSS	no DSA performed	n.d.	n.d.	bradykinesia, gait disturbances and resting tremor of upper extremities	3 yrs	n.d.	none	Madopar	died of seizures and pulmonary infection
18	Fujii [13], 2014	69/M	SSS	thrombus in SSS	supplied by the superficial temporal artery and occipital artery; retrograde flow into the cortical veins in the frontal and parietal lobes	Borden type II	cognitive dysfunction and parkinsonism	2 yrs	none	the latter part of the SSS was occluded; TV embolization using platinum coils of the venous sinus at the shunting point → n.d./no angiography, unclear if complete occlusion was achieved	L-dopa 300 mg daily	improved
19	Ma [14], 2015	62/M	SSS	n.d.	cDAVF in L temporal region, fed by bilateral middle meningeal arteries and meningeal branches of vertebral artery which were enlarged abnormally, with poor venous reflux to SSS	n.d.	parkinsonism and progressive memory loss	5 mo.	bilateral frontal lobes	endovascular embolization; near-complete occlusion; recurrence L temporal DVA at discharge	anti-parkinson therapy	improved
20	Kim [15], 2015	75/M	L TSS	n.d.	L MMA to L transverse and sigmoid sinuses; retrograde venous sinus drainage	n.d.	severe bradykinesia and rigidity involving the axial muscles and all 4 limbs; short-stepped gait with decreased bilateral arm swing and mild postural instability; both hands exhibited resting tremor (4–5 Hz)	4 mo.	thalamus, globus pallidus, and cerebellum	endovascular embolization; → complete occlusion was achieved	no anti-parkinson therapy	improved

Table 1 (continued)

Case No.	First author [ref.], year	Age, yrs /sex	Fistula location	cDAVF etiology – risk factors	cDAVF anatomy (feeding vessels, venous reflux)	cDAVF classification	Neurological symptoms	Time from onset to treatment	Hyperintense lesions on T2-weighted image	Therapeutic intervention	Medication	Follow-up overall outcome
21	Pu [16], 2017	51/M	straight sinus	straight sinus occlusion	cDAVF adjacent to the tentorial notch fed by the PMA, meningohypophyseal trunk, and a branch of the left internal carotid artery; the cortical venous drainage drained into the vein of Galen, which flowed in a retrograde direction into the vein of Rosenthal, as well as into the internal cerebral veins	Cognard grade IIa grade IIb	remarkable hypomimia, slow speech, hypophonia, psychomotor slowness, generalized bradykinesia and brisk deep tendon reflexes; rigidity in all 4 limbs, with some hypokinesia on rapid alternating movements; mild weakness in the lower limbs	1 mo.	medial part of lenticular nuclei bilaterally and frontal lobe white matter	endovascular embolization through the L complete artery → complete occlusion was achieved	Madopar 62.5 mg 3× daily	improved
22	Lai [17], 2017	62/M	cDAVF involving SSS, both transverse sinuses, torcula, and R sigmoid sinus	a history of unprovoked TSS thrombosis and unprovoked lower extremity DV thrombosis	diffuse retrograde cortical venous drainage and reflux into the DV system	n.d.	bradykinetic, hypomimia, hypophonia, and symmetric cogwheeling rigidity	2 mo.	hyperintensity in cerebral hemispheric white matter	TV Onyx and coil embolization of the torcula and R TSS fistula, with minimal residual fistulous flow → recurrence of fistula → resection of the involved part of the SSS with fistula disconnection, and repeat TV Onyx embolization of the residual R transverse sinus fistula via direct puncture of the fistula through a burr-hole → complete occlusion was achieved	L-dopa 400 mg daily	improved
23	Lai [17], 2017	65/F	R TSS	R sigmoid sinus occlusion, L transverse sinus stenosis	reflux into the DV system	n.d.	L appendicular ataxia, atypical bilateral coarse hand and arm tremor, bradykinesia, rigidity, and generalized stimulus-induced myoclonus	3 mo. to 5 yrs	hyperintensity in cerebral and cerebellar white matter	TV coil embolization of the fistula → complete occlusion was achieved	none	improved

Table 1 (continued)

Case No.	First author [ref.], year	Age, yrs /sex	Fistula location	cDAVF etiology – risk factors	cDAVF anatomy (feeding vessels, venous reflux)	cDAVF classification	Neurological symptoms	Time from onset to treatment	Hyperintense lesions on T2-weighted image	Therapeutic intervention	Medication	Follow-up overall outcome
24	Chang [18], 2019	57/M	L transverse-sinus	thrombosis of the L sigmoid sinus	early opacification of the L transverse sinus with occlusion of the sigmoid sinus; reverse blood flow to the straight sinus and R transverse sinus, and many engorged cortical veins	n.d.	L forearm showed rigidity and bradykinesia; it was difficult for the patient to smoothly perform rapid alternating movement testing	n.d.	diffusion-weighted image showed high signal intensity above right basal ganglion, internal capsule, and thalamus	TV embolization failed; a combined surgical and endovascular approach was performed with puncturing of the L transverse sinus directly with an 18-G needle sheath through a burr-hole in the L occipital bone and insertion of a microcatheter into the L transverse sinus via the sheath; detachable coils were deployed in the distal sigmoid sinus posterior to the distal L transverse sinus → after embolization, only minimal residual fistula with slow flow remained → 1-month follow-up DSA showed complete fistula occlusion	none	improved
25	Our study	57/M	straight sinus	thrombosis of the straight sinus	the fistula was fed by the tentorial dural arteries and drained through the median tentorial sinus into the culmen vein, superior vermillion vein and then the system of the vein of Galen; due to the thrombosis of the proximal segment of the straight sinus and missing direct outflow, the fistula caused reflux and congestion in the system of the internal cerebral veins.	Cognard type III Borden type III DES-Zurich BVS with direct, exclusive, and strained LVD	psychomotor slowdown and cognitive impairment with disorientation; small-stepped gait, rigor of all extremities, reduced oscillation of the arms, and hypomimia	4.5 mo.	venous congestive edema of both thalami, internal capsule, hippocampi, pallidum, and mesencephalon predominantly on the R side due to the non-occluded cDAVF and multiple microhemorrhages within the congested tissue	several attempts at venous embolization were unsuccessful; microsurgical fistula clipping with disconnection of the culmen vein from the medial tentorial sinus was performed → complete occlusion was achieved	Madopar + ropinirole	improved

L, left; R, right; M, male; F, female; BVS, bridging vein shunt; DSA, digital subtraction angiography; MMA, middle meningeal artery; PMA, posterior MA; TAE, transarterial embolization; DVA, developmental venous anomaly; DV, deep venous; TV, transvenous; TSS, transverse-sigmoid sinus; SSS, superior sagittal sinus; n.d., no data; mo., month(s); yrs, years; wks, weeks.

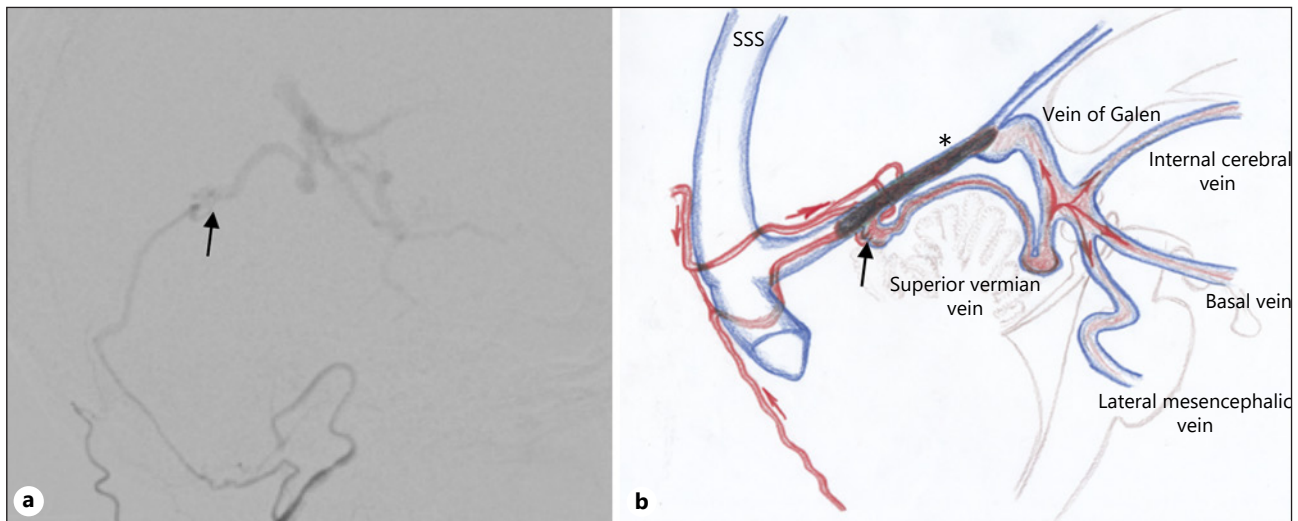


Fig. 1. **a** DSA reveals an arteriovenous shunt (arrow) at the level of the thrombosed part of the straight sinus. **b** Graphic illustration of the cDAVF. The fistula is fed by the tentorial dural arteries (red) and drains through the straight sinus into the median tentorial sinus, supraculminar vein, superior vermian vein, and then into the system of the vein of Galen. Due to the thrombosis (asterisk) of the proximal segment of the straight sinus, missing direct outflow, and arterialization of the system of the vein of Galen with consecutive flow reversion, the fistula causes severe reflux and congestion in the system of the internal cerebral veins. The cDAVF is classified as Cognard type III, Borden type III, and DES-Zurich bridging vein shunt type with direct, exclusive, and strained LVD. SSS, superior sagittal sinus.

icant medical history. After unspecific cranial computed tomography (CT) and cerebrospinal fluid examination, cranial magnetic resonance imaging (MRI) revealed partial thrombosis of the straight sinus with diffuse edema of both thalami, the internal capsule, the hippocampi, the pallidum, and the base of the mesencephalon (right > left) as well as multiple microbleeds in the basal ganglia and dilated deep cerebral veins.

Digital subtraction angiography (DSA) was performed, which showed a typical DAVF at the level of the thrombosed part of the straight sinus. The fistula was fed by the tentorial dural arteries and drained through the straight sinus into the median tentorial sinus, supraculminar vein, superior vermian vein, and the system of the vein of Galen. Due to the thrombosis of the proximal segment of the straight sinus, missing direct outflow, and arterialization of the system of the vein of Galen with consecutive flow reversion, the fistula caused severe reflux and congestion in the system of the internal cerebral veins (Fig. 1a, b). It was thus classified as Cognard type III, Borden type III, and DES-Zurich BVS type with direct, exclusive, and strained LVD (Fig. 1).

At an external hospital, several attempts at venous embolization of the fistula were not successful. Transarterial embolization was not performed due to high-grade outlet stenosis of the left vertebral artery. A transarterial embolization attempt via the transosseous-supplied posterior meningeal artery did not seem promising and was therefore not attempted. The patient was referred to our department 6 weeks after initial presentation.

At the time of admission, the patient showed severe neurological impairment and was not able to take care of himself, requiring considerable assistance and frequent medical care (Karnofsky performance score [KPS] of 30).

Repeated MRI and magnetic resonance angiography (MRA) revealed a massive progression of the venous congestive edema of both thalami, the internal capsule, the hippo-

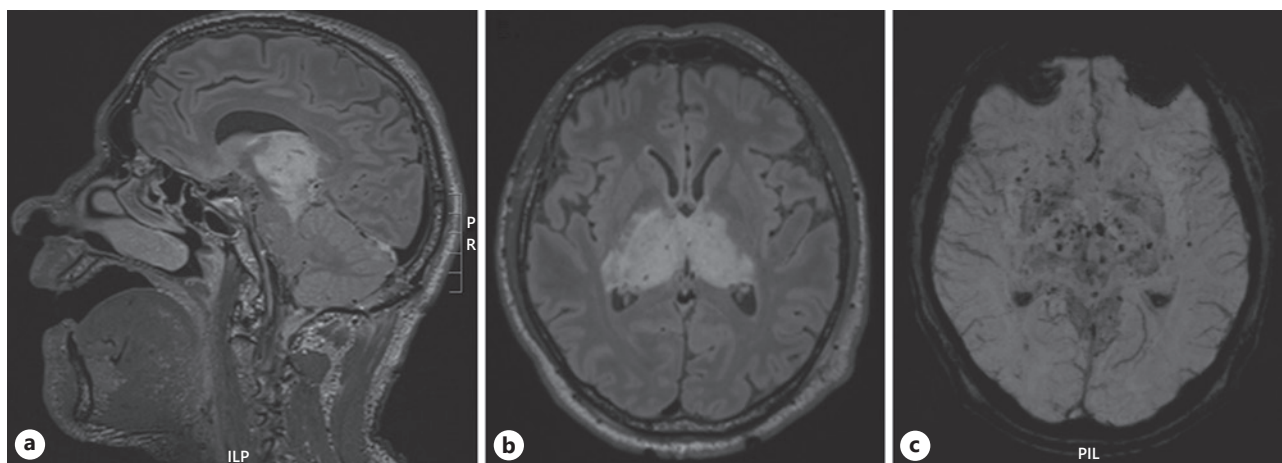


Fig. 2. **a, b** Sagittal and axial section of FLAIR-weighted images show a massive progression of the venous congestive edema of both thalami, internal capsule, pallidum and mesencephalon due to the nonoccluded cDAVF. **c** SWI-weighted images show multiple microhemorrhages within the congested tissue.

campi, the pallidum, and the mesencephalon, predominantly on the right side, due to the nonoccluded cDAVF (Fig. 2a, b), and also multiple microhemorrhages within the congested tissue (Fig. 2c).

A suboccipital paramedian right-sided mini-craniotomy was performed. Microsurgical opening of the cisterna magna was performed to facilitate supracerebellar infratentorial access and to identify the fistula draining into the supraculминаl and superior vermian veins. Verification of its arterialization was done by fluorescein angiography. The culминаl vein was disconnected from the medial tentorial sinus by bipolar electrocoagulation, achieving an immediate fistula occlusion. Verification of the successful disconnection was also done by fluorescein angiography. Postoperative MRA and DSA showed complete obliteration of the cDAVF (Fig. 3b, d). During hospitalization, a detailed examination of thrombophilia was performed due to the unexplained thrombosis of the straight sinus. There was no evidence of thrombophilia or oncological disease. At discharge, 12 days after surgery, the patient showed a significantly improved general condition. He was, however, not able to walk by himself and his cognitive impairment was still severe (a KPS of 60).

Carbidopa/levodopa was started based on the diagnosis of a secondary akinetic rigid Parkinson syndrome, and the neurological condition of the patient further improved. In the further course of the treatment, the dose was increased and ropinirole was added so that the patient was free of Parkinson symptoms when he left the rehabilitation clinic.

Three months later, the patient presented at our outpatient clinic in a significantly improved clinical condition (a KPS of 80–90). There were no signs of parkinsonism and his cognitive condition had improved. MRI showed complete regression of the bithalamic edema, internal capsule, hippocampi, pallidum, and mesencephalon (Fig. 4b, d), although the thrombosis of the straight sinus had remained unchanged.

Due to the complete regression of the venous stasis edema and the working diagnosis of a secondary Parkinson syndrome, dopaminergic medication was slowly tapered with no subsequent deterioration of the patient observed.

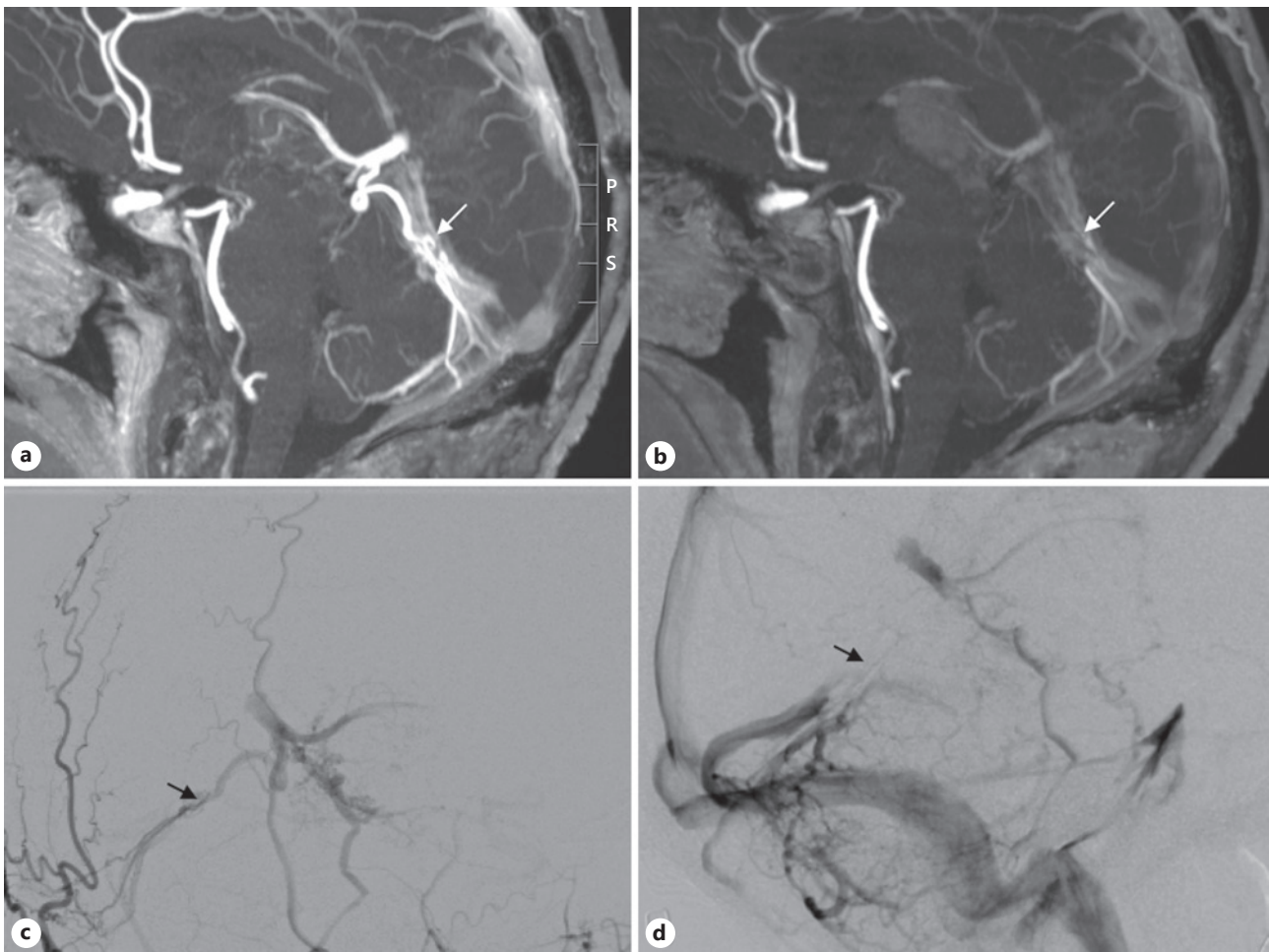


Fig. 3. Sagittal section of T1-weighted MRI TOF with gadolinium before (a) and after microsurgical clipping and complete cDAVF occlusion (b). DSA before (c) and after complete cDAVF occlusion (d).

Discussion

We present here the case of a 57-year-old male with a cDAVF of the straight sinus (Borden type III, Cognard type III, DES – Zurich BVS-type with direct, exclusive, and strained LVD) and worsening venous congestion of the Galenic system. He presented clinically with executive dysfunction, cognitive impairment, and disorientation. After complete microsurgical fistula occlusion, the bithalamic edema (with involvement of the internal capsule, hippocampi, and mesencephalon) regressed. Concomitantly, parkinsonism disappeared and his functional impairment improved significantly.

Parkinsonism as a clinical manifestation of cDAVF is exceptionally rare and has so far only been reported in case reports and small case series [27]. We identified 24 cases described as having cDAVF and associated parkinsonism in the literature (Table 1). AV shunting into the transverse and sigmoid sinuses ($n = 14$; 58.3%) was most often described, followed by the superior sagittal sinus ($n = 3$; 12.5%), the torcular, straight sinus, and multiple sinus involvement (each $n = 2$; 8.3%), and anterior cranial fossa ($n = 1$; 4.2%). Retrograde filling of the straight sinus and/or impaired drainage of the deep internal venous system were

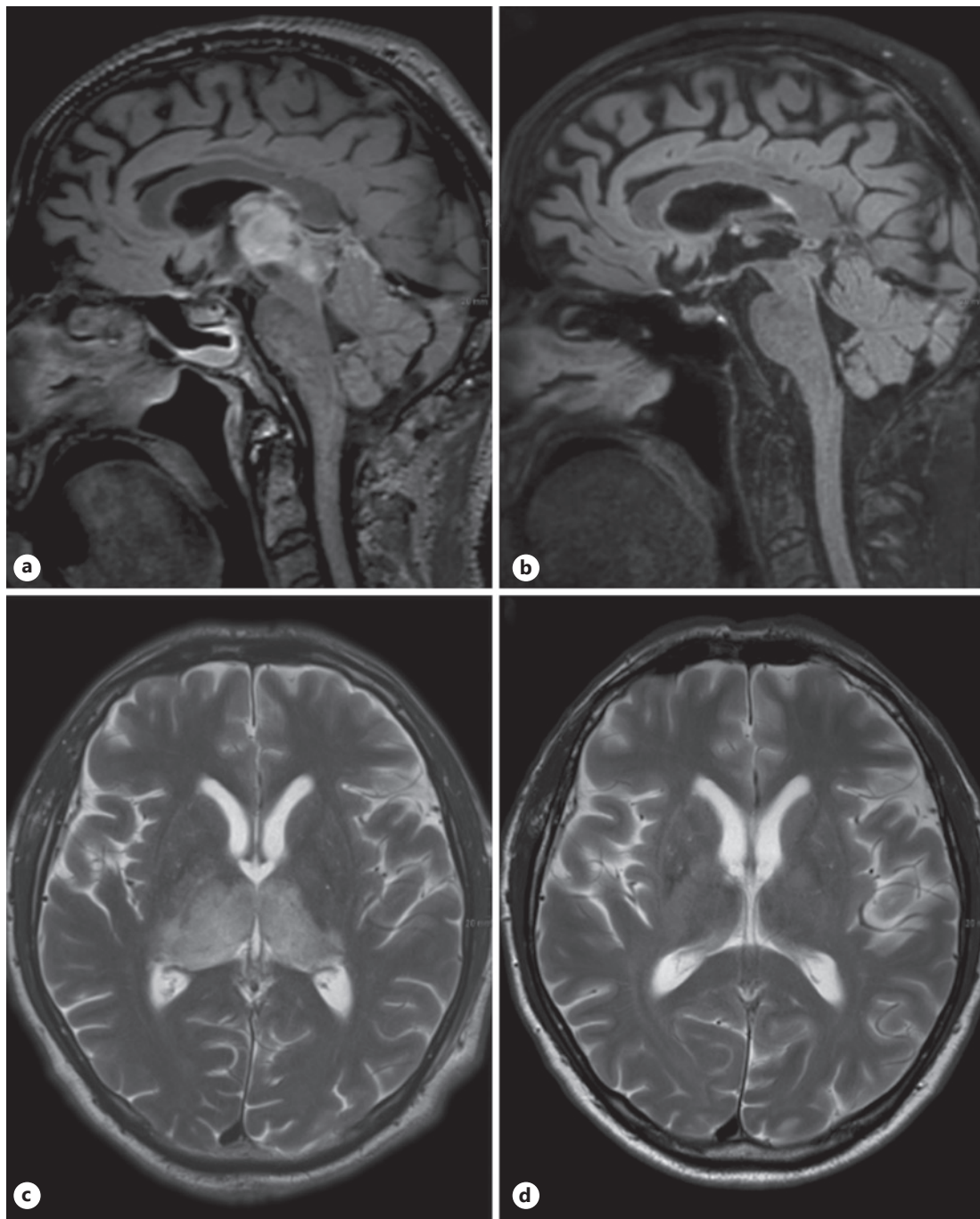


Fig. 4. Sagittal section of FLAIR (**a, b**) and axial section of T2-weighted (**c, d**) MRI. The 3-month follow-up MRI displays complete regression of the venous congestion of the Galenic system (**b, d**) compared to preoperatively (**a, b**).

described in 16 patients (66.6%). Limited data or no data on the venous reflux were provided for 8 patients (33.3%).

Lee et al. [20] were the first to propose the theory of basal ganglia dysfunction due to impaired drainage of the deep internal veins in patients with cDAVF and parkinsonism. They

described a 60-year-old patient with cDAVF of the left sigmoid sinus with retrograde filling of the deep venous system, and superior sagittal sinus with prominent cortical venous reflux [20]. The patient showed reversible parkinsonism after embolization, which was well correlated with an increase in the basal ganglia-cerebellar perfusion ratio on SPECT, suggesting that a perfusion defect was responsible for the pathogenesis of the parkinsonism [20].

Kim et al. [15] reported a patient with decreased ^{18}F -FP-CIT uptake in cDAVF-associated parkinsonism. They suggested that hemodynamic impairment could cause parkinsonism via an accentuation of the underlying dopamine deficiency in subjects with preclinical-stage parkinsonism. The parkinsonism in their patient improved following the endovascular occlusion of the cDAVF without antiparkinson drug therapy [15].

In our case, both the presynaptic dopaminergic neurons in the mesencephalon and the postsynaptic neurons in the striatum, essential to mediate the treatment effect of dopaminergic medication, were affected by the edema (Fig. 2a, b). In addition, given the fact that carbidopa/levodopa was started during the process of neurological improvement, it remains unclear if the first signs of improvement during rehabilitation were related to this dopaminergic medication or to surgery. We believe it is more likely that the microsurgical disconnection of the cDAVF was primarily responsible for the clinical improvement during rehabilitation. The medication could therefore be tapered later without symptom recurrence. In the literature review, it was found that dopaminergic medication did not lead to improvement in 4 patients [13, 17, 20, 24] and that a transient response was reported in 3 patients [12, 16].

Treatment of patients with cDAVF and LVD in the deep venous system with secondary parkinsonism and/or cognitive impairment is challenging, as a remission of symptoms has been described in only a few cases in which complete fistula occlusion was achieved. The literature review illustrates the severe natural history of cDAVF; 2 patients died (no intervention), and 3 patients showed severe neurological deterioration (1 had no intervention and 2 attempted unsuccessful embolization). These numbers might even be underestimated due to limited data on follow-up in the cases described and the known general bias against reporting poor results [9, 11, 12, 24].

Our aim is to highlight the importance of rapid diagnosis and complete occlusion of the cDAVF to reverse parkinsonism and prevent further neurological deterioration, including death. Interestingly, complete cDAVF occlusion was achieved in only 8 of 24 (33.3%) and incomplete occlusion in 5 patients (20.8%). In 3 patients (12.5%), no intervention was made; insufficient data were available for 8/24 (33.3%) (Table 1). In all the reported cases, endovascular embolization was the 1st treatment choice. In 3 cases, no intervention was performed; in 1 case, no information was available about the treatment used. Surprisingly, no cDAVF in this literature review was treated by microsurgical cDAVF disconnection, despite complete occlusion being achieved in only 8/24 (33.3%) patients. Endovascular embolization was repeated in a few cases when the first attempt was unsuccessful (Table 1). In 3 cases, a combined endovascular and surgical approach was performed to gain access to the cDAVF for endovascular treatment, but microsurgical clipping/disconnection of the cDAVF was not performed in these cases (Table 1) [17, 18, 23].

Even in cases where only incomplete cDAVF occlusion was achieved through endovascular embolization, a “watch-and-wait-strategy” was the preferred strategy, exposing the patient to persistent LVD and the risk of clinical deterioration.

The detailed understanding of the angioarchitecture of the cDAVF is essential to determine the treatment strategy. It is based on the precise identification of the AV shunt localization (BVS, DSS, or EVS) as well as on the understanding of the LVD. A detailed review of the angioarchitecture of cDAVF is beyond the scope of this report and we refer the interested reader to the description by Baltsavias and Valavanis [1–3, 6]. To achieve complete cDAVF occlusion, a multidisciplinary discussion of the endovascular and microsurgical options is recom-

mended. Although we favor endovascular surgery as the first-line treatment, microsurgery should be considered in cases of persistence of an AV shunt. The surgical approach is often straightforward and can be easily performed by an experienced vascular neurosurgeon. Despite a preoperatively detailed understanding of the angioarchitecture of the cDAVF, intraoperative identification of the AV shunt might pose a challenge; confirmation of the AV shunt as well as the successful disconnection should therefore be verified by intraoperative fluorescein angiography [28].

Patients with cDAVF diagnosed as BVS, the majority of which are Borden type III, are excellent candidates for primary microsurgical cDAVF occlusion [29]. The surgical approach is straightforward and can be done through a simple mini-craniotomy by simply disconnecting the draining vein, regardless of the presence or absence of venous strain [29].

Statement of Ethics

The subject gave his written informed consent to publish this case. In addition, the institutional review board approved the use of registry data for clinical research, registered under the case No. KEK-ZH 2012-0244.

Conflict of Interest Statement

None of the authors report a conflict of interest related to this report.

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Author Contributions

J.V., Z.K., and L.R.: conception and design; acquisition, analysis, and interpretation of data; drafting the article. F.B. and H.R.: acquisition, analysis, and interpretation of data.

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